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CASE REPORT

Necklace calcification on chest radiographs in infants in cardiac intensive care

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Introduction

Soft-tissue calcification is a recognized feature of many conditions, often with characteristic patterns and distribution.[1] We describe four children with severe congenital cardiac disease, who developed fine, bilateral soft-tissue calcification in the root of the neck and around the shoulder girdle soon after an episode of acute shock and hypoxia. Similar calcification has been previously described in case reports, and in each it has occurred after an episode of severe shock or hypoxia, though with different antecedent causes. These appearances seem to be linked to tissue hypoperfusion and hypoxia, and are likely to be due to brown fat necrosis.

Case reports

Case 1

A 1-week-old term infant presented with lethargy and jaundice. He was found to have poor left ventricular function, was hypoxic and acidotic (pH 7.0, serum lactate 7.12 mmol/l (normal range 0.63–2.44)) and required ventilatory support. He required peritoneal dialysis for deteriorating renal function 5 days after admission. Ten days after admission, fine calcification in the soft tissues of his neck was noted on chest radiography (Fig. 1). This calcification became more extensive over subsequent weeks, and extended over the chest wall.

The patient's condition deteriorated and he died 2 months after admission. His serum calcium was slightly elevated on admission (2.99 mmol/l (normal range 2.25–2.80)) but normal thereafter. Serum phosphate was elevated (peak 2.77 mmol/l (normal range 1.30–2.00)) for the first 5 days of admission. There was no history of soft-tissue trauma or extravasation of fluid at the site of the calcification.

Post-mortem examination demonstrated myocardial infarction due to fibromuscular dysplasia of the left anterior descending artery. Calcification was identified in the adipose tissue around the heart and in the subcutaneous tissues of the thorax and neck. The coronary vessels were not calcified.

Case 2

An 8-day-old term male infant was admitted with hypotension and acidosis. Clinical and echocardiographic examination demonstrated an interrupted aortic arch and an aorto-pulmonary window. On admission his renal function was normal, but it deteriorated rapidly and he required peritoneal dialysis.

He remained hypoxic (pO_2 45 mmHg) and unstable, and required continuing ventilatory and inotropic support. His interrupted aortic arch was repaired 5 days after admission, after which his condition improved, and within a month of his surgery his renal function returned to normal and he was extubated.

One month after surgery, fine, bilateral soft-tissue calcification was noted around the base of the neck on chest radiographs (Fig. 2). This was not present on previous radiographs and was still present on discharge. His serum calcium levels (corrected) had been 2.08 mmol/l on admission and 2.94 mmol/l on the day of surgery. Otherwise they were within normal limits. He had received 2 days of intra-venous calcium gluconate on admission, and none thereafter. His phosphate levels were transiently elevated (4.33 mmol/l) on admission, but normalized quickly and remained so thereafter. There was no extravasation of fluids, trauma or cutaneous abnormalities at the site of the calcification. He was subsequently discharged from hospital having made a good recovery. His calcium and phosphate levels remained normal.

Case 3

A 4-week-old boy was admitted electively for aortic valvotomy and mitral annuloplasty. The surgery was uncomplicated but post-operatively he developed respiratory syncytial virus (RSV) pneumonitis and required ventilation. His condition deteriorated and he developed bilateral pneumothoraces, and ultimately required extra-corporeal membrane oxygenation (ECMO) support. During this time he was hypotensive, and he developed acute renal failure, which required peritoneal dialysis. His renal function had been normal on the day of admission. He was noted

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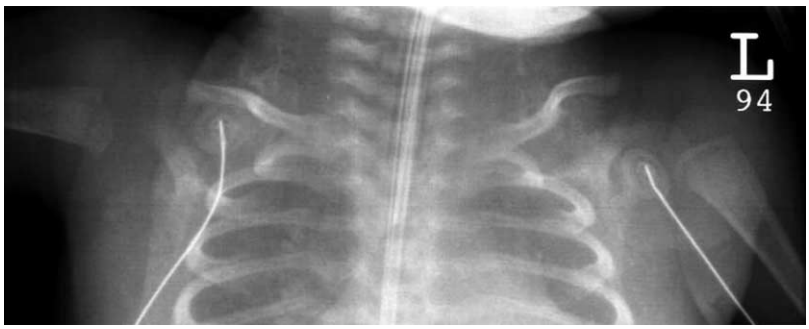

Figure 1 There is calcification bilaterally in the root of the neck in a "necklace" distribution.

to have post-operative mitral regurgitation and had mitral valve replacement 2 months after his original operation.

After 1 month of peritoneal dialysis, he developed fine bilateral soft-tissue calcification in the root of his neck, which had not been present previously (Fig. 3). His serum calcium levels were within normal limits, except on one occasion (1.94 mmol/l) 1 month before the calcification was noted. His phosphate levels were intermittently elevated (maximum 2.33 mmol/l). There was no history of soft-tissue trauma, extravasation of fluid or cutaneous abnormality. He made a good recovery and was discharged from hospital.

Case 4

A female infant (gestational age 31 weeks) was delivered prematurely at 31 weeks gestation because of maternal pre-eclamptic toxaemia and gestational diabetes. The infant was noted to be hypoxic (90%), tachycardic, tachypnoeic and hypotensive (mean arterial pressure 34 mmHg). She was intubated on day 1 but remained hypoxic and hypotensive, and developed congestive cardiac failure.

Echocardiogram demonstrated truncus arteriosus and a patent ductus arteriosus. Her condition stabilized and she underwent definitive surgery at the age of 2 months.

During the procedure she developed recurring ventricular tachyarrythmias and closure of the chest was delayed. She developed renal failure in the immediate post-operative period and required 8 days of peritoneal dialysis, after which she made a good recovery.

Fine bilateral calcification was noted in the soft tissues of the root of her neck on a chest radiograph 9 days after the operation. This persisted for 3 months before disappearing. The serum calcium level was normal throughout, other than on the day of surgery (2.96 mmol/l). The serum phosphate was elevated (3.29 mmol/l) 3 days post-operatively, and then quickly returned to normal. Both serum calcium and phosphate had normalized at out patient review.

There had been no trauma, extravasation of fluids or cutaneous abnormality at the site of the calcification.

Discussion

The patients presented all developed a similar pattern of soft-tissue calcification, in similar clinical settings. They were all acutely unwell with a cardiac problem, and each was hypoxic and hypotensive, requiring ventilation and peritoneal dialysis for acute renal failure. Their renal function was normal on admission. There was no history of injury or extravasation of fluids at the site of the calcification in any of the patients. Each had minor fluctuations in levels of serum calcium and phosphorus but no underlying metabolic abnormality.

Soft-tissue calcification has many differing causes and can be broadly divided into three main groups. Calcification may occur in normal tissues in the presence of abnormally high serum calcium or

Figure 2 Fine, bilateral calcification again present in root of neck.

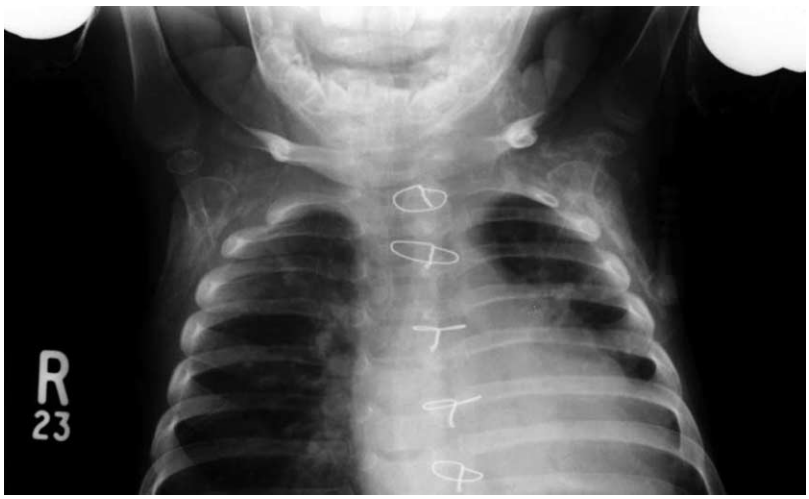

Figure 3 Similar bilateral neck calcification again noted.

phosphate levels (elevated calcium–phosphorus ion product). It may occur in abnormal or damaged tissue even when calcium and phosphorus biochemistry is normal—dystrophic calcification. It may also occur in the presence of normal tissues and biochemistry.[1]

Soft-tissue calcification may occur in patients with chronic renal failure due to secondary hyperparathyroidism. These patients tend to have persistently high phosphate levels with normal calcium levels, causing the calcium–phosphorus ion product to be increased. The calcification in these patients tends to be peri-articular, although soft-tissue, visceral, and blood vessel calcification is also common.

The patients presented had acute renal failure, not chronic, and the calcification appeared within 1 month of the initial renal insult. There was also no evidence of any peri-articular calcification, as seen in the shoulders and hips during multiple chest and abdominal radiographs. The calcification, which was noted during the post-mortem, was in fat only with no vessel calcification present.

Calcification in these cases is therefore unlikely to be due to the renal failure, which was present in each case, given the distribution of the calcification and the lack of chronicity in these cases.

Subcutaneous and perivisceral brown fat is normally present in the soft tissues of infants, including the cervical area. Brown fat is very vascular and plays an important role in homeostasis, especially temperature control. It is, therefore, particularly vulnerable to vascular hypoperfusion, as in shock, and brown fat necrosis can result. Dystrophic calcification then may occur, making it radiologically visible.

Soft-tissue calcification in brown fat necrosis has been previously described in a critically ill infant with hypoxia secondary to broncho-pulmonary dysplasia.[2] In this case, the calcification was mainly axillary, with extension into the neck, and in the peri-renal areas. There has also been a post-mortem study of 400 babies,[3] in which 22 of the children had brown fat necrosis. In this series only peri-adrenal and peri-tracheal fat was studied, but it was felt that the cause of the necrosis was shock. Most of these babies died from so-called sudden death syndrome, but three had congenital heart disease. The occurrence of the fat necrosis did not offer any clues as to the cause of the shock.

Soft-tissue calcification in the neck in infants has also been described in a child who was a victim of child abuse. This child was admitted for the second time after a prolonged spell of apnoea, for which she needed resuscitation. On the second occasion, her chest radiograph revealed fine, necklace distribution calcification, along with healing rib fractures. She had documented hypoxic ischaemic encephalopathy, and it was postulated that the calcification was due to strangulation. However, this was not proven, and as in the present cases, the child had suffered hypoxia within weeks of developing the calcification.[4,5] Moreover, the calcification extended towards the axillae, which may suggest hypoxia rather than direct trauma as the cause in this case.

Fine, necklace calcification in the neck seems to occur, and become radiographically visible, in infants after severe shock or hypoxia. This seems likely to be due to brown fat necrosis. It is not a marker of prognosis or of the underlying cause of the shock or hypoxia.

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